A CASE OF PERONEAL MUSCULAR ATROPHY (TYPE CHARCOT-MARIE).

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The following case is worthy of record as a clinical example of the type of peroneal muscular atrophy.

The patient, an intelligent, unmarried man, aged 42 years, gives a very clear history of his trouble, and of his family.

The family history is entirely negative, except concerning the father, of whom the patient remembers nothing, though he has never been told that his father had any symptoms like his own. An only sister has five healthy children. His mother died of an acute intestinal disorder. A half-brother of his father died of pulmonary tuberculosis.

The patient is 6 feet 3 inches tall, and weighs 156 lbs. He has never weighed more than 165 lbs. He walks with some difficulty. His gait is distinctly that of "steppage," owing to bilateral foot-drop. While standing he constantly changes the position of his feet (treading) in order to maintain his equilibrium, using his eyes at the same time as an aid. He cannot stand still and talk with a person without the aid of a cane or other support. At night he feels very uncertain in walking. He is left-handed but uses his right hand in writing. The grip in each hand is very powerful, but there is clearly a slight degree of incoördination in the hands; for he buttons his clothing with difficulty, and coarse coördinated movements of the arms show some slight lack of precision. He is not aware of any trouble in his hands or arms.

The face and tongue present no peculiarities. The eyes are in all respects normal save for a slight degree of presbyopia of recent date. All the higher senses are normal. There are no anomalies of the vegetative organs, and his general health is excellent.

Of late years the patient has found occupation in a country store. As a school teacher he became unable to move about with required promptness. Before teaching school he did farm work.

He was healthy until the age of 16, and then had an attack of acute articular rheumatism; one joint after another became swollen and painful, and he was ill for six weeks.

About fifteen years ago he noticed the gradual development of numerous, very painful corns on both feet. To avoid pain he tried walking in various ways, and finally some twelve years ago, after suffering with corns for a long time, he began to have shoes made to prevent suffering. At this time he noticed that his toes had become flexed, this causing prominence of the joints. He cannot recall the beginning of the weakness and atrophy now so noticeable in his legs, for as he states, both have developed so gradually. Aside from the pain due to corns, he has never had any kind of pain in his legs, though occasionally he has cramps in the calves of his legs at night, so severe that he has to get up and walk about to get relief.

He has never been a drinker, but has chewed tobacco to excess for years. He has never had any venereal disease. Five years ago he had pneumonia, and an attack of malaria three years ago.

During the last five years he has noticed a decided weakening of sexual power; lack of desire and infrequency and deficiency of erection. At no time has there been any vesical or rectal disturbance.

He states that he has seen, and more frequently felt, twitchings in the muscles of his legs; but this on the whole was rare and has not been noticed of late.

As already stated, the gait is that of "steppage" with a slight, though very evident, degree of incoördination. Romberg's sign is very marked. Without assistance, the patient would fall in an attempt to stand with the eyes closed.

Careful examination of the sensory functions reveals no disturbance anywhere of senses of touch, pain, and temperature. With his eyes closed, the patient is perfectly aware of the position of all his limbs, and he instantly and correctly indicates any passive movements communicated to arms, legs, hands, feet, fingers or toes.

Voluntary movement is everywhere normal, except in the legs and feet. He can move the left foot slightly in dorsal flexion. He can extend the right great toe slightly. All other voluntary movements of the feet and toes are impossible.

The power of the muscles of the thighs is normal, save for some slight weakness of the flexors of the knee on each side. When leaning forward, the patient has some trouble in bringing the trunk to an erect position on the pelvis. The muscles of the buttocks are soft and flabby, and evidently weak though not reduced in volume when compared with the musculature in general. Flexion of the thighs on the pelvis is normal in force. In the upper extremities there is nothing abnormal in movement, but there is distinct weakness of the right deltoid, without atrophy.

Fibrillary twitching is noticeable in the outer heads of each gastrocnemius muscle close to the knee as the patient stands.

The spinal column is markedly prominent in the lower dorsal and lumbar regions; a general and equal prominence without an angle.

There is marked vasomotor disturbance in the legs. The thighs are warm as low as the beginning of the lower third, but from this point downwards the limbs are cold, and the feet are bluish and a little edematous. The patient has a sensation of coldness of feet and legs, and goes to great trouble in dressing to keep them warm.

The toe-nails are much thickened and corrugated. He has no corns at the present time.

There is no tenderness of muscles or nerve-trunks anywhere.

Muscular atrophy is far advanced in all the muscles of both legs, and is quite symmetrical; it is noticeable in the thighs up to the beginning of the middle third. The vasomotor anomaly has a distribution exactly corresponding with the distribution of muscular atrophy.

The deformity due to the muscular atrophy is double talipes equino-varus of moderate degree. There is retraction shown in the flexed and elevated position of the toes, but the feet are "balottants," when the leg is passively shaken.

All the superficial reflexes are present and lively. The plantar reflex is remarkably lively, with flexion (slight) of the toes, and retraction of the whole limb (both sides). The deep reflexes at the ankles are absent. The knee-jerks are both present. That on the right side is very slight without reinforcement; that on the left side appears feebly with Jendrassik's maneuver. The reflexes of the upper extremities are normal.

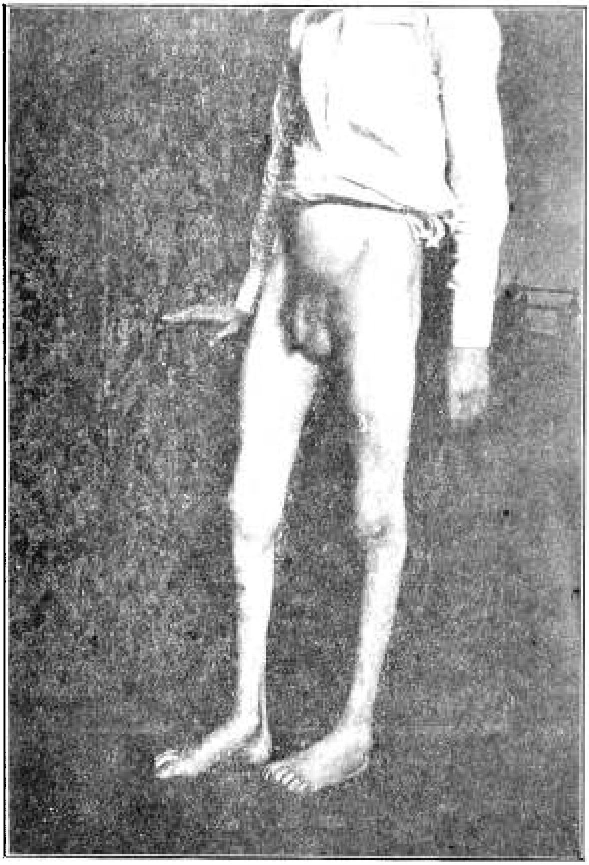
All the muscles of the thighs, even the atrophied recti, respond normally to faradism and galvanism. In the muscles of the legs and feet no response to either can be obtained. Elsewhere electrical reactions are normal.

The accompanying reproduction of a photograph shows clearly the areas affected by the muscular atrophy.

The history of this case shows that the trouble began about the age of 24; that probably the peroneals and extensors of feet and toes were first attacked (corns due to flexion of toes). Doubtless the weakness and atrophy of the muscles of the legs developed *pari passu*, and came on so insidiously and slowly that years elapsed before the patient knew to what to attribute his increasing difficulty in walking, deceived at first by the assumption that it was entirely due to corns.

The family etiological element is entirely wanting. Acute rheumatism eight years before the trouble was noticed is worthy of emphasis.

The case corresponds in all points with cases of amyotrophy of the Charcot-Marie type, but there is one symptom present, the absence of which is emphasized in all cases thus far reported, and which I have not seen in any case that has come under my observation, *i.e.*, the marked and unmistake-



The photograph shows the areas affected by the muscular atrophy.

able Romberg's sign. If, as is held by some observers, the posterior columns of the cord are degenerated in this disease, the usual absence of Romberg's sign calls for explanation.

In this case there is some evidence that other muscles are destined to be invaded, as shown by the condition of the glutei and the right deltoid. The right thenar muscles also present a suspicion of atrophy. The slight incoördination of the hands and arms is also noteworthy.

The weakening of sexual power in this patient is a symptom to which I have found no reference in recorded cases of this disease.

It should be added in conclusion that the patient has always lived in a very low malarial district near the Mississippi.